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Short communication

Neonatal eating epilepsy: 50-year follow-up

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ABSTRACT

Purpose: We present a case of neonatal eating epilepsy.
Method: Case report.
Results: An otherwise healthy female newborn started having epileptic seizures lasting up to 5 minutes induced by breastfeeding at age of 2 weeks. The convulsive seizure started with crying and coughing followed by opisthotonus, flexion of upper extremities, extension of lower extremities, and generalized jerks in extremities. The patient had fairly often postictal vomitings which could not be relieved by upright positioning. At baseline clinical examination, no abnormal symptoms or signs were found except for a postictal hoarseness. On the day of admission to the tertiary care hospital, during 1.5 hours, she had 10 attacks. Though some attacks were spontaneous and during sleep, most were induced by feeding or — less often and inconsistently — by manipulating the pharynx. Phenobarbital 15 mg four times a day stopped the attacks. The last seizure was documented 14 days after admission and phenobarbitalwas eventually withdrawn. The patient remained seizure free off AEDs during a prospective follow-up for over 50 years until 2013.
Conclusions: Neonatal eating epilepsy may be a suitable diagnosis if convulsive seizures are precipitated by feeding or eating. Polygraphic recording will help ascertain the diagnosis. Antiepileptic therapy is likely to abolish seizures.

1. Introduction

Eating epilepsy is a rare epilepsy syndrome in which all seizures are precipitated by sensory stimulation of pharynx and/or larynx, e.g., associated with feeding or eating.[1] The typical age at onset of eating epilepsy seems to range from 8 to 55 years.[2–6] To our knowledge, only three cases of neonatal eating epilepsy have been reported albeit with a much shorter follow up.[7–9] In our prospectively followed population sample of 245 children who met the criteria for epilepsy i.e. two or more unprovoked seizures[10] and identified from the catchment area of the Turku University Hospital from 1 January 1961 to 31 December 1964,[11–15] 1 (0.41%) patient was identified as a case of eating epilepsy which began in the neonatal period and was followed up for over 50 years.

2. Case description

The girl was born after uneventful pregnancy at term in good health as the fourth child to the non-sanguinous parents in 1961. No epilepsy or other nervous system diseases were detected in the family history. Except for mild edema during the last trimester, pregnancy and delivery were uneventful. The birth weight was 3080 g.

At age 2 weeks, she began to present with attacks associated with the beginning of breast feeding. The semiology was similar in all attacks. They started with crying and coughing followed by opisthotonus, flexion of upper extremities, extension of lower extremities, and generalized jerks in extremities. Foaming spittle was noted. Weakening breathing was interrupted by vigorous breathing followed by apnea associated with dark blue skin coloration. Mouth-to-mouth resuscitation and oxygen via a mask were given and the attacks stopped within about 5 min. The patient had fairly often postictal vomitings which could not be relieved by upright positioning. At baseline clinical examination, no abnormal symptoms or signs were found except for a postictal hoarseness. On the day of admission to the tertiary care hospital, during 1.5 h, she had 10 attacks. Though some attacks were spontaneous and during sleep, most were induced by feeding or—less often and inconsistently—by manipulating the pharynx. Interictal clinical and X-ray examination showed no abnormality in heart or lung function. Kine-X ray investigation with contrast media revealed no pharyngo-esophageal dysfunction. In the evening of the day of hospitalization, there was a severe, unprovoked attack with a short period of cardiac

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arrest. In an interictal EEG recording, conducted after discontinued phenobarbital medication, asymmetric background activity of lower amplitude and repeated slow-wave discharges of several seconds on the right side compared with the left one were found.

Phenobarbital 15 mg four times a day and chlorpromazine 3 mg three times a day, started immediately after hospitalization, stopped the attacks. Administration of chlorpromazine was discontinued after six days. Fast tapering of phenobarbital prior to EEG recording one week after admission to the hospital was associated with seizures. Following an increase of the daily dosage only few seizures were seen. The last seizure was documented 14 days after admission. After four weeks of in-patient treatment, the patient was discharged with phenobarbital 10 mg twice daily. Weight gain was normal during the hospitalization.

During post-discharge follow-up, the patient was in remission and the medication was slowly withdrawn without any seizure relapse. The health status has been prospectively followed for more than 50 years until 2013. Except for heavy pollen and other allergies, the patient remained in good health. She is employed, and lives a normal life. During a follow-up program, a 3T MRI investigation (without exposure to contrast media because of a prior allergic reaction) revealed no abnormal brain pathology.

3. Discussion

We report the long-term follow-up of a patient with neonatal eating epilepsy presenting with convulsive seizures that were associated with feeding. Treatment with phenobarbital was successful. Tapering of the medication resulted in seizure recurrence that was stopped after going back to the original phenobarbital dose. The patient was followed up for 52 years and remained seizure-free off AEDs.

Eating epilepsy with onset during the neonatal period is very rare. To our knowledge, there are three reported cases of neonatal eating epilepsy with a much shorter follow up of 4–14 months.[7–9] An infant with benign familial neonatal convulsions developed hemiclonic and multifocal clonic seizures lasting 1–2 min. The majority occurred immediately after breast feeds.[7] Some were preceded by version of the eyes and head and asymmetric tonic posturing. At the start of the seizure, the EEG showed a build-up of high-voltage slow-wave activity in the right anterior temporal region, which then spread to the left and became generalized. No treatment was given and the seizures ceased spontaneously on day 10. He has since developed normally, as followed up to the age of 14 months. His mother had identical episodes from days l to 10, which were ascribed to colic, without any problems since.[7] A further case of neonatal eating epilepsy started at day 1 with 2–3 episodes per day of cyanosis and increased muscle tone lasting 1 min and induced by breast feeding.[8] The interictal EEG showed right central–temporal epileptiform discharges. Treatment with phenobarbital stopped the episodes, but the EEG was not improved until antacids were administered. A diagnosis of severe GER was made based on abnormal esophageal pH monitoring with a follow-up of 12 months. There was no family history of seizures and the MRI was normal.[8] The third case is a 5 months old child whose eyes would turn to left (partial seizure), followed by cessation of sucking, choking and cyanosis, and atonic seizure.[9] The whole episode would last for less than a minute. After this the child would sleep for 1 h and was back to normal at waking up. These episodes would occur mainly during breast feeding, while sucking and sometimes after breast feeding was finished. The episodes had a frequency of 2–3 times per day and each episode lasted a maximum of twenty seconds. The frequency increased on the last day to 5 times in association with cyanosis and lip smacking. Many times mother noted chocking like phenomenon while sucking at breast. There was no history of regurgitation or vomiting. The girl was born normally after an uneventful pregnancy. There were no neonatal problems. Her development was consistent with her age. There was a strong family history of seizures. Four cousins on maternal side had seizures. Three of them were off medication now while the fourth one continued to have active seizures and was on treatment. One uncle and aunt on fathers side also had seizures in the past and were off medications now.[9] Routine EEG and MRI were normal. There was right posterior rhythmic temporal slowing while breast feeding. The child was started on sodium valproate at 10 mg/kg/day in two doses which stopped the seizures. She remained seizure free for the last four months on 80 mg two times a day (20 mg/kg/day). She was developing as a normal child.[9]

The typical age at onset of eating epilepsy beyond the neonatal cases described seems to range from 8 to 55 years.[2–6] Eating epilepsy was found in only 9 (0.045%) of 20,000 patients investigated in an EEG laboratory in Italy[16] and in 13 (0.11%) of 11,783 outpatients in Japan.[2] A review of the files of about 9000 outpatients with epilepsy in Turkey revealed 6 (0.067%) cases of eating epilepsy.[17] Striking exceptions are Asian countries, particularly. Epilepsy seems to be more common in Asia, particularly in Sri Lanka. A prospective study of 1287 patients with epilepsy in Sri Lanka revealed 191 (14.8%) patients whose seizures were associated with eating.[18] The higher prevalence rate and clustering in families has been suggested to be associated with bulky meals rich in carbohydrate.[2,3,19] The definition of eating epilepsy, however, may be broader as interpreted in the Asian studies. Eating epilepsy included all patients "who had more than 50% of fits during or within 30 min of eating breakfast, lunch or dinner".[3] Wieser et al.[20] studied the time between the stimulus and reaction and suggested that latencies exceeding several minutes do not warrant the diagnosis of reflex epilepsy in its strict sense.

Our single case report cannot and does not intend to contribute to elucidate the putative mechanism(s) of neonatal feeding-induced seizures and neonatal eating reflex epilepsy.[21] However, several regions of the brain have been implicated in the literature to being involved in initiating neonatal feeding-induced seizures. These include the diencephalon,[22] the hypothalamus[23] and the amygdala.[24,25] Although our case with several feeding-induced seizures meets the criteria of reflex epilepsy,[1,26] the nosological entity of neonatal reflex eating epilepsy has been challenged in the literature (see[21]). Given the apparent heterogeneity of the eating epilepsies, Loiseau et al.[27] preferred to speak of eating-induced seizures.

There are several conditions which must be considered in the differential diagnosis of neonatal eating epilepsy. Gastroesophageal reflux (GER) disease in infants typically presents with regurgitation and vomiting.[28] The attacks of our patient were not typically those of vomiting and weight gain was normal. GER is reportedly not linked to apnea.[29,30] Sandifer syndrome, first described in 1964,[31] includes a symptom complex of GER, hiatal hernia, odd body posturing and torticollis.[32,33] Werlin et al.[34] described a 16-day-old girl who had postprandial episodes of shaking, body stiffening, and hyperextension of both arms followed by regurgitations. EEG was normal. Thickened feedings and upright positioning stopped vomiting. Our case had an abnormal EEG, bilateral jerks between the feedings, and the quality of food or feeding position did not help stop attacks. Hyperekplexia may include tonic stiffening attacks with cardiac asystole precipitated by feeding or defecation. There are, however, typical other triggers like sudden loud noises, choking, sneezing and bath water, and the symptomatology includes sudden startles, frequent harlequin color changes, and antiepileptic medications have no satisfactory effect.[35] Epileptic seizures may occur as apnea and seizures and GER in extremely rare cases even as comorbidities. Navelet et al.[36] reported a 3-month-old boy who presented with repeated episodes of cyanosis, apnea, bradycardia and

hyperactive movements of the upper limbs, first diagnosed as esophagitis, but then ascertained by polygraphy as epileptic seizures and followed within some few months by severe epilepsy with tonic-clonic seizures. Finally, eating seizures have been reported to broadly include movement-induced motor seizures triggered by chewing.[27]

Several findings argue in favor of our diagnosis of neonatal eating epilepsy as a neonatal reflex epilepsy. They include seizure induction by sucking or feeding, absence of typical GER symptomatology, no relief from upright positioning, presence of convulsions, exclusion of GER and, most importantly, a clearcut effect of AED therapy. The medical and social outcome of our patient is very favorable with life-long seizure freedom off AEDs after neonatal events.

In conclusion, neonatal eating epilepsy may be a suitable diagnosis if convulsive or complex partial seizures are precipitated by sucking, feeding or eating. Polygraphic recording will help ascertain the diagnosis. Antiepileptic therapy is likely to abolish seizures.

Conflict of interest

This study was not sponsored. Dr. Sillanpää and Dr. Schmidt report no financial disclosures.

References

1. Engel JJ. A proposed diagnostic scheme for people with epileptic seizures and with epilepsy: report of the ILAE Task Force on classification and terminology. *Epilepsia* 2001;**42**:796–803.
2. Nagaraja D, Chand RP. Eating epilepsy. *Clin Neurol Neurosurg* 1984;**86**:95–9.
3. Senanayake N. "Eating epilepsy" – a reappraisal. *Epilepsy Res* 1990;**5**:74–9.
4. Nakazava C, Fujimoto S, Watanabe M, Tanaka M, Ishikawa T, Watanabe Y. Eating epilepsy characterized by periodic spasms. *Neuropediatrics* 2002;**33**:294–7.
5. Seneviratne U, Seetha T, Pathirana R, Rajapakse P. High prevalence of eating epilepsy in Sri Lanka. *Seizure* 2003;**12**:604–5.
6. Patel M, Satishchandra P, Saini J, Bharath RD, Sinha S. Eating epilepsy: phenotype, MRI, SPECT and video-EEG-observations. *Epilepsy Res* 2013. http://dx.doi.org/10.1016/j.epilepsyres.08.05 (Epub ahead of print).
7. Baxter P, Kandler R. Benign familial neonatal convulsions: abnormal intrauterine movements, provocation by feeding and ICTAL EEG. *Seizure* 1997;**6**:485–6.
8. Domizio R, Conte E, Puglielli C, Domizio S, Maragni S, Pollice R, et al. Neonatal eating epilepsy: pathophysiological and pharmacologic aspects. *Int J Immunopathol Pharmacol* 2006;**19**:697–702.
9. Koul R, Aida AlShihi A, Mani R, Javad H, AlFutaisi A. Eating epilepsy or feeding epilepsy in an infant. *Eur J Paediatr Neurol* 2013. http://dx.doi.org/10.1016/j.ejpn.2013.10.004.
10. Commission on Classification and Terminology. Proposal for revised classification of epilepsies and epileptic syndromes. *Epilepsia* 1989;**30**:389–99.
11. Sillanpää M. Medico-social prognosis of children with epilepsy. Epidemiological study and analysis of 245 patients. *Acta Paediatr Scand* 1973;**237**(Suppl. 68):1–104.
12. Sillanpää M, Jalava M, Kaleva O, Shinnar S. Long-term prognosis of seizures with onset in childhood. *N Engl J Med* 1998;**338**:1715–22.
13. Sillanpää M, Shinnar S. Status epilepticus in a population-based cohort with childhood-onset epilepsy in Finland. *Ann Neurol* 2002;**52**:303–10.
14. Sillanpää M, Haataja L, Shinnar S. Perceived impact of childhood-onset epilepsy on quality of life as an adult. *Epilepsia* 2004;**45**(8):971–7.
15. Sillanpää M, Shinnar S. Long-term mortality in childhood-onset epilepsy. *N Engl J Med* 2010;**363**:2522–9.
16. Vizioli R. The problem of human reflex epilepsy and the possible role of masked epileptic factors. *Epilepsia* 1962;**3**:293–302.
17. Kokes U, Baykan B, Bebek N, Gurses C, Gokyigit A. Eating epilepsy is associated with initial precipitating events and therapy resistance. *Clin EEG Neurosci* 2013;**44**:161–6.
18. Senanayake N. Reflex epilepsies: experience in Sri Lanka. *Ceylon MJ* 1994;**39**:67–74.
19. Mandal DK, Bandyopadhyay S, Tarafdar J, Mukherjee A, Chaudhury S, Deb A, et al. Eating epilepsy: a study iof twenty cases. *J Indian Med Assoc* 1992;**90**:9–11.
20. Wieser HG, Hungerbühler H, Siegel AM, Buck A. Musicogenic epilepsy: review of the literature and case report with ictal single photon emission tomography. *Epilepsia* 1997;**38**:200–7.
21. Wolf P, Koepp M. Reflex epilepsies. *Handb Clin Neurol* 2012;**107**:257–76.
22. Robertson J, Fariello WCRG. Eating epilepsy associated with a deep forebrain glioma. *Ann Neurol* 1979;**6**:271–3.
23. Sepulveda FC, Duro LA, Da Silva MN, Leite SR. Epileptic crisis induced by food intake: report of a case. *Ass Neuropsiquiatr* 1981;**39**:106–14.
24. Ahuja GK, Mohandas S, Narayanaswamy AS. Eating epilepsy. *Epilepsia* 1980;**21**:85–9.
25. Fiol ME, Leppik IE, Pretzel K. Eating-epilepsy: EEG and clinical study. *Epilepsia* 1986;**27**:441–5.
26. Fisher RS, Acevedo C, Arzimanoglou A, Bogacz A, Cross JH, Elger C, et al. An operational clinical definition of epilepsy. *Epilepsia* 2014. http://dx.doi.org/10.1111/epi.12550.
27. Loiseau P, Guyot M, Loiseau H, Rougier A, Desbordes P. Eating epilepsy. *Epilepsia* 1986;**27**:161–3.
28. Czinn SJ, Blanchard S. Gastroesophageal reflux disease in neonates and infants: when and how to treat. *Paediatr Drugs* 2013;**15**:19–27.
29. Jeffery HE, Ramilly P, Read DJC. Multiple causes of asphyxia in infants at high risk for sudden infant death. *Arch Dis Childh* 1983;**58**:92–100.
30. Walsh JK, Farrell MK, Keenan WJ, Lucas M, Kramer M. Gastroesophageal reflux in infants: relation to apnea. *J Pediatr* 1981;**99**:197–201.
31. Kinsbourne M, Oxon DM. Hiatus hernia with contortions of the neck. *Lancet* 1964;**1**:1058–61.
32. Gellis SS, Feingold M. Syndrome of hiatus hernia, with torsion spasms and abnormal posturing. *Am J Dis Child* 1971;**121**:43–54.
33. Bray PF, Herbst JJ, Johnson DG, Book LS, Ziter PA, Condon VR. Childhood gastrointestinal reflux. *JAMA* 1977;**237**:1342–5.
34. Werlin SL, D'Souza BJ, Hogan WJ, Dodds WJ, Arndorfer RC. Sandifer syndrome: an unappreciated clinical entity. *Dev Med Child Neurol* 1980;**22**:374–8.
35. Ferrie CD, Stephenson J, Rees MI, Parsons J, Livingston J. Hyperekplexia with recurrent prolonged cardiac asystole. *Dev Med Child Neurol* 1998;**39**(Suppl. 77):5–6.
36. Navelet Y, Wood C, Robleux C, Tardieu M. Seizures presenting as apnoea. *Arch Dis Childh* 1989;**64**:357–9.